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Case report

Solitary multilocular cyst of the kidney — a diagnostic problem

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Benign multilocular cystic nephroma is a rare condition of the kidney, first described in the literature by Edmunds in 1892.[1] The terminology is a confusing issue and the name has many synonyms, the most popular being multilocular cyst of the kidney. Preoperatively this diagnosis is difficult to make even with today's sophisticated diagnostic techniques, and this case highlights that problem.

CASE HISTORY. A 58-year-old woman presented after having taken an overdose of paracetamol. During routine physical examination a non-tender mass was noted in the right outer quadrant of her abdomen. She previously had surgery for repair of a perforated duodenal ulcer, a left mastectomy for infiltrating ductal carcinoma, and a tympanoplasty for cholesteatoma. She also suffered from depressive neurosis and diverticular disease of the colon. She had no family history of renal disorder. She was normotensive and routine blood investigations were normal. Mid-stream urine sample revealed asymptomatic infection with *Escherichia coli.*

Ultrasound scan revealed a 6 × 7 cm cystic lesion in the lower pole of the right kidney. Some distortion of the mid and lower pole calyceal systems was noted on intravenous urography; the left renal tract was normal. CT scan revealed a mixed cystic and solid lesion of the right kidney, a distended gall bladder, a dilated common bile duct and compression of the duodenum. There was loss of the tissue plane between the structures, giving an impression of invasion (Fig 1). Barium meal demonstrated irregularity of the second and third part of the duodenum in keeping with an invasive nature of the tumour observed on CT scanning.

The initial diagnosis after these investigations was of a malignant renal neoplasm; no secondary spread was demonstrable on chest X-ray, liver or bone scan. Operation was performed through a right paramedian transperitoneal

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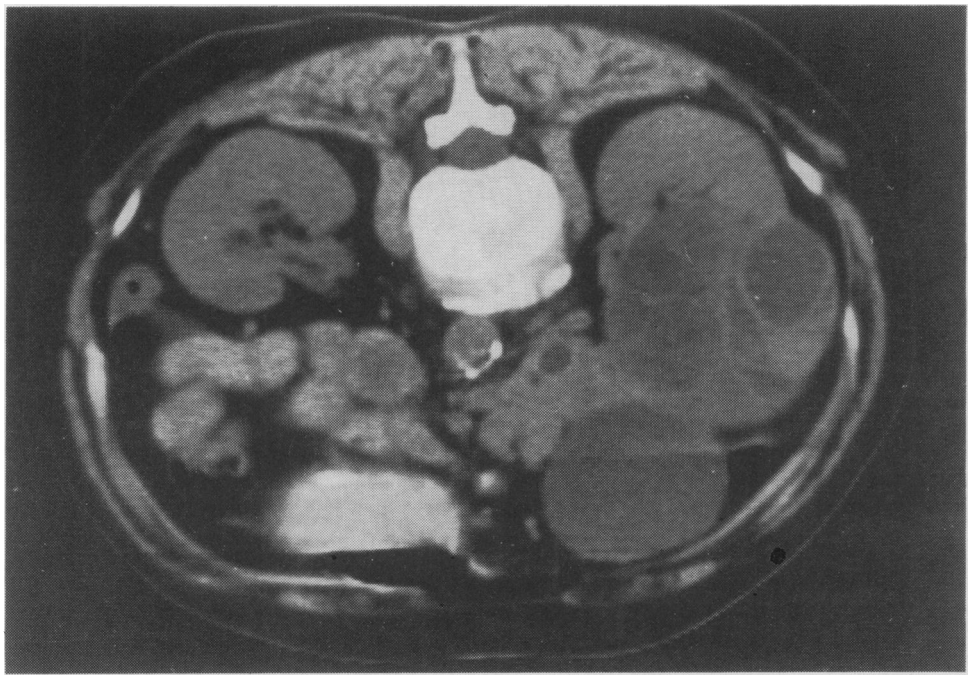

Fig 1. CT scan showing a large cystic lesion attached to the right kidney and suggesting invasion of neighbouring structures.

approach. The lesion was a well circumscribed cystic mass and there was no evidence of invasion of the adjacent organs. Routine nephrectomy was performed. The 13 × 8 × 5 cm kidney weighed 470 gm. A well circumscribed multiloculated cystic lesion occupied the lower pole, impinging on the calyceal system. The cut surface revealed many locules filled with straw coloured fluid. There was no communication with the pelvis (Fig 2). Histologically the lesion comprised individual cysts lined by flat attenuated epithelium. The septae between the cysts were composed of nondescript cellular mesenchyme and hyalinised collagenous tissue. There was a distinct surrounding fibrous capsule which separated the lesion from the underlying compressed renal parenchyma. The renal vein and ureter were normal.

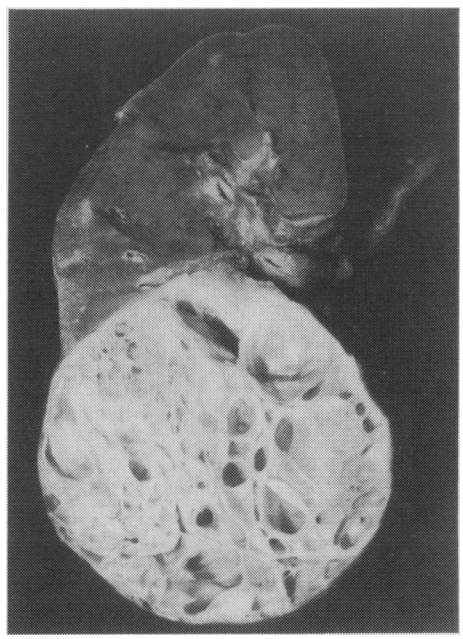

Fig 2. Well delineated multiloculated cyst occupying one pole of the right kidney.

DISCUSSION

Benign multilocular cystic nephroma, although a rare entity in the literature, is now more often recognised. The diagnosis is made using the eight criteria set down by Powell et al,[2] later modified by Boggs and Kimmelstiel.[3] These are — 1) the lesion should be unilateral, 2) it should be solitary, 3) it should be multilocular, 4) the cyst should not communicate with the renal pelvis, 5) the loculi should not communicate with one another, 6) the loculi should be lined with epithelium, 7) fully developed nephrons should be absent from the septa of the cyst and 8) the remaining kidney tissue should be normal. This case satisfies all these criteria. The terms used for this tumour are many and include cystadenoma, cystic renal harmatoma, polycystic Wilms'tumour and cystic nephroma.

The exact pathogenesis is unknown, various authors proposing different theories. Meland and Braasch[4] support a congenital or developmental origin, whereas Christ[5] suggests a neoplastic one. Baldauf[6] claims a multifactorial cause. Others consider it to be a harmatoma[7,8] while many like Boggs and Kimmelstiel[3] believe that it is a benign neoplasia arising from metanephric blastema. The commonest mode of presentation is with a mass in the abdomen, but patients may present with pain, haematuria or infection. There are no systemic effects associated with the lesion, in contrast to polycystic kidneys, although there have been cases of patients with hypertension becoming normotensive following removal of the lesion.[7] Total nephrectomy is commonly resorted to because the lesion is difficult to diagnose preoperatively.[9,10] Ultrasound and CT scans are helpful[7,10] but as demonstrated here, are by no means completely reliable. Simple deroofing may be followed by a recurrence,[11] but partial nephrectomy has been performed with success.[9]

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